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Discussion

Dr Richard A. Jonas (Washington, DC). I have no disclosures.

Dr Barron, many thanks for sending me the manuscript. This is obviously a complex area and you have a huge amount of data, so it was very helpful to be able to look at the manuscript ahead of time.

You are certainly to be congratulated on this additional, extremely important contribution regarding the challenging problem of ccTGA. It was just over 20 years ago that Dr Michel Ilbawi introduced the concept of the double switch (DS) procedure and held out hope for this difficult problem of ccTGA. Until that time the results of traditional surgery, which left the right ventricle as the systemic ventricle, resulted in dismal long-term outcomes, as was so ably documented by Dr Viktor Hraska when he was with our group in Boston. Although encouraging early results were reported for the DS by Professor Imai's group at Tokyo Women's Medical College and Dr Roger Mee in Melbourne, as well as Dr Brawn and your group in Birmingham, this most recent analysis

will surely lead all of us to rethink which patients are best managed by this procedure.

I would like to highlight some of the disturbing points that Dr Barron has just presented to us. First, late left ventricular dysfunction is not uncommon. Recent reports had already raised that suspicion, but what is of greater concern is that late left ventricular dysfunction cannot be accurately predicted. I think we were all hopeful that training of the left ventricle at an older age, such as greater than 12 or 15 years, or the presence of preoperative heart failure or the presence of postoperative valve dysfunction might have been predictive of late left ventricular dysfunction. Regrettably, none of this has been documented by the analysis we have just heard. In fact, this large experience raises the question whether the fundamental premise that the child with ccTGA must always be better off with a systemic mLv and a biventricular circulation is correct.

What is of equal concern to the high incidence of late left ventricular dysfunction is the high incidence of late need for reoperation and reintervention. The DS patients not only had a higher incidence of late left ventricular dysfunction relative to the RS patients, but in addition they had a surprisingly high rate of late aortic valve regurgitation and pulmonary artery problems, particularly after a Lecompte maneuver. These problems were less frequently seen in the RS patients, but surely this latter option is in other respects a less desirable option than a true DS since it condemns patients to a lifetime of conduit changes. Conduit changes can be a particular problem when the patient has dextrocardia, which applied to more than 20% of your patients in this series and in many other series has applied to as many as one third of all patients. And for both groups reoperations were needed for the Senning pathways, both systemic and pulmonary, which leads to my questions:

First, is there ever a place for a prophylactic DS in the child with an intact septum and no TR? What sort of associated problems like a mild degree of Ebstein-like malformation of the TV or congenital heart block might lead you to do a prophylactic DS?

Dr Barron. Thank you very much for your very fair comments. I completely agree. It has raised concerns in all our minds about the long-term management of these patients. Absolutely, it is patient selection—who needs what operation at what time—that is the key.

I am not sure whether many people would ever have suggested that doing a prophylactic DS in a child who is completely well, has no TR, and a good functioning systemic right ventricle, that putting them through this major maze operation, would be the right thing to do. So no, I do not think there is any place for prophylactic DS.

Now, if they have a degree of TR, then it starts to get a bit trickier because the natural history is bad. As soon as they have moderate or greater TR, they are on a bad survival curve, and that is more difficult. The first thing I would say is that I think there is a place for a PAB in that situation, because that in itself may treat the TR and buy some time. You are setting them on that track toward DS at that stage. However, that may well be their best outcome despite what we are saying here.

Dr Jonas. Would it be fair to say that the bar is shifted slightly against the DS in terms of these patients who do not have a VSD and who have an intact septum?

Dr Barron. Yes, I think so. That's a fair comment. Of course, we are all conscious of the patients in whom symptoms develop. The more difficult problem is the ones who do not have symptoms but whose right ventricle does not look good or who have some TR.

Dr Jonas. Second, how important is dextrocardia as a contraindication to an RS procedure in view of the risk of proximal conduit compression by the sternum and need for frequent conduit changes?

Dr Barron. In our series I would not say it has been a major concern, and we certainly would not regard it as a contraindication to the DS. Certainly dextrocardia makes the operation technically more difficult to do, but we can be reassured by the fact that the operative results for the RS have been very good. If you look at the incidence, we have only had to actually replace 12 conduits in the whole population over the 19 years of this study. So the commitment to lots of operations for conduit replacement does not seem to be too bad.

We have not analyzed it specifically to look at whether the dextrocardia patients are those who have needed more conduit replacement, so I could not answer that.

Dr Jonas. Third, I have a question about PAB. To whom do you now offer left ventricular preparation by PAB? Do you see a role for PAB as destination therapy to induce septal shift to treat TR in the patient with an intact septum and low left ventricular pressure?

Dr Barron. I do not think we would consider PAB unless the patients had evidence that they were running into trouble, that they had moderate TR, or that they had some dysfunction or some signs of dilatation of the mRV. Those are the patients we would select for banding at that point. I quite agree that the banding itself will cause septal shift and may reduce the TR and symptoms in many of the patients.

Whether that will be the destination, I do not think I could answer. I think each patient must be evaluated individually. Once PAB is done, you are making a commitment that you would be expecting to take them through a DS. If they remain well with the PAB on, they should be safe and there is no reason not to just keep watching them, indefinitely if need be. But in our experience, with the majority of patients, once TR and right ventricular dilatation have developed, they are going to ultimately get systemic ventricular failure and they will need something.

Dr Jonas. You can certainly make the argument that the left ventricle is going to manage the banded pressure over the longer-term. If the TR is stable and the septum remains shifted across, perhaps in the long-term that would be a more satisfactory outcome than some of the multiple reinterventions that you have documented.

Final question: What is the role of the Fontan operation versus the DS in light of the results that you have presented?

Dr Barron. There are certain populations within this group of patients for whom the Fontan would be a good and a valid alternative. I do not think there is any role for it in the DS-type population, because you have not removed the concern of the mRV failing or the TR becoming a problem. If you take them through a Fontan program, that risk is still there.

I guess the Fontan is more appropriate in the group of patients who are in the RS group, in whom you would avoid the whole problem of having to place a conduit, perhaps the risk of heart block and the risk of leaving a left ventricle-aortic tunnel. And you know that the Fontan is a pretty safe operation.

We are all kind of hard-wired to think that if the patients have 2 ventricles, it is a sin to take them through a Fontan circulation, and that is something we can debate. However, I think the functional performance and the functional result the patients get with a biventricular repair, which in this case means RS versus the Fontan, is better with an RS. Thus I would still put my vote for a biventricular repair with an RS, and I think that is supported by these outcomes. We know that the Fontan, although safe, has a long-term attrition. I would argue it might be worse than we are going to see in the RS group. Certainly there are patients in the RS group who have an uncommittable VSD, and in those patients I think there is a very good argument for doing a Fontan rather than a conventional physiologic repair.

Dr Jonas. Thank you. Once again, congratulations, this is going to be a really important contribution.

Dr Rodolfo A. Neirotti (*Cambridge, Mass*). Congratulations for your large number of patients and for being honest in presenting the data in which you have shown some long-term problems that can be useful for others trying to use the same approach.

I would like to expand on Richard Jonas's question, and that is, the place of the Fontan operation. In patients with ccTGA and large VSDs, the ventricular dysfunction appears usually after the septation. In this cohort of patients, after the Fontan operation, the 2 ventricles function as a large single chamber, with a single outlet, an anatomy and physiology that may result in a better ventricular function than when you do the septation and the DS.

Dr Barron. No, I think that is a fair comment. If you use the Fontan approach, they will fall into the better series of Fontan outcomes. However, I still believe that a biventricular repair provides a better functional result. The results here would support the fact that so long as they have a suitable anatomy where the Rastelli is readily performed, the left ventricle can be committed back to the aorta without causing any obstruction within the ventricle. I think the functional outcome for those patients would still be superior than to do a Fontan.

Dr V. S. Reddy. David, I think you can discuss it later because we are way over time.